Case Report

Acute Pulmonary Edema due to Rupture of Mitral Valve´s Posteromedial Papillary Muscle in a young girl

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ABSTRACT

We report a 13-year-old girl with acute pulmonary edema due to severe acute Mitral regurgitation because of rupture of Mitral valve´s posteromedial papillary muscle. She had normal mitral valve with necrotic posteromedial papillary muscle, classic aneurysms in left coronary artery, neutrophilic infiltration in the wall of medium-sized vessel of kidney and positive perinuclear antineutrophil cytoplasmic antibodies (P-ANCA) a diagnosis of polyarteritis nodosa. She treated successfully with mitral valve replacement. (Rawal Med J 2008;33:112-114).

Key Words: Mitral valve, P-ANCA, Papillary muscle, polyarteritis nodosa.

INTRODUCTION

Microscopic polyarteritis nodosa is a vasculitis syndrome similar to Wegener's granulomatosis. Microscopic polyarteritis nodosa as with Wegener's granulomatosis, shows arteritis in the medium to small arteries and microangiitis especially in the lung and kidney.[1] There has been no case report of microscopic polyarteritis nodosa complicated with severe acute mitral regurgitation due to papillary muscle rupture.[1-3] Papillary muscle rupture is rare but catastrophic complication in cardiology practice. In this condition with detiorating hemodynamics, early diagnosis and immediate cardiopulmonary support are required before surgical treatment.[4-5] This report presents a case of Microscopic polyarteritis nodosa complicated by rupture of mitral valve´s posteromedial papillary muscle and acute pulmonary edema that was treated successfully with surgical mitral valve replacement.

PRESENTATION OF CASE

A 13-year-old girl was admitted to Madani Heart Center, Tabriz, Iran because of acute pulmonary edema. She had intermittent arthralgia, which affected mainly the left wrist. Six days before admission, vomiting developed in the evening. She reported that she had no fever, chills, diarrhea, cough, anorexia, weight loss, or rash and that she had not been exposed to ill persons or ingested unusual foods. On physical

examination, the heart rate was 161 and the systolic blood pressure was 90 mm Hg first and then dropped to 70 mm Hg. The extremities were cool. Prominent crackles were heard over the chest. There was a grade 3 holosystolic apical murmur, without a thrill. Examination of the abdomen revealed no abnormalities. No peripheral edema was seen. No rash, petechiae, ecchymoses, oral lesions, or lymphadenopathy was found. The arms and legs were normal. Neurologic examination revealed no abnormalities. The urinary sediment contained 25 to 30 red cells, 1 to 3 white cells, no red-cell casts, and a few bacteria per high power field. Some laboratory data are shown in Table 1.

Table 1. Laboratory data.

Variable	**Result**
Hematocrit (%)	30.3
White blood cell Count(/m3)	19600
Neutrophils (%)	83
Lymphocytes (%)	7
Monocytes (%)	5
Eosinophils (%)	5
Platelet Count(/m3)	236000
Reticlulocyte (%)	5.1
Erythrocyte sedimentation rate (mm/h)	125
Prothrombine time (sec)	13.5
Partial thromboplatine time (sec)	35
Urea nitrogen	26
Creatinine	1.3
Blood Sugar (mg/dl)	93
Na^+ (mmol/liter)	132
K^+(mmol/liter)	3.7
Aspartate aminotransferase(U/liter)	57

An electrocardiogram showed a sinus tachycardia at a rate of 115, with nonspecific ST-segment and T-wave abnormalities. A chest radiograph revealed diffuse, bilateral air-space disease consistent with the presence of pulmonary edema. The oxygen saturation was 76 percent while the patient was breathing ambient air. Oxygen by face mask and Furosemide were administered, but cardiopulmonary collapse ensued. She was intubated and urgently transferred to intensive cardiac care unit.

Transthoracic echocardiography revealed severe acute mitral regurgitation, normal left ventricular size with reduced function and inferior wall hypokinesia. A transesophageal echocardiographic examination showed marked prolapse of the middle and medial scallops of the anterior mitral-valve leaflet (Fig 1 and 2). There was failure of coaptation of the central scallops of the mitral valve, resulting in severe mitral regurgitation that was directed posterolaterally. There was no evidence of flail leaflet, ruptured chordae, or vegetations. The valve leaflets were neither thickened nor myxomatous. The left atrium was dilated; the left ventricular size was normal, but systolic function was mildly impaired, and there was a subtle, inferior wall-motion abnormality associated with the abnormal papillary muscle. The proximal epicardial coronary arteries appeared normal. All other findings were normal.

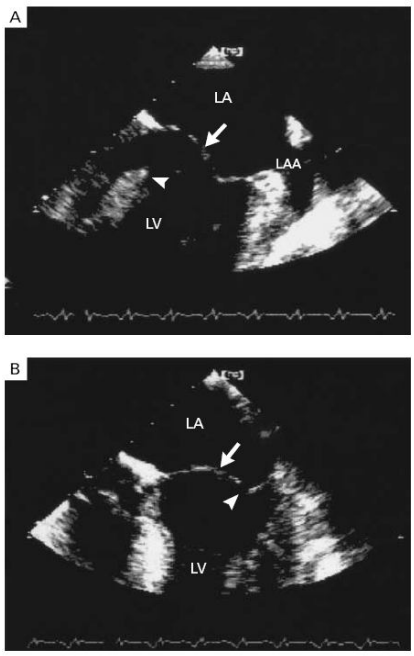

Figure 1. Transesophageal Echocardiogram.
The view in Panel A shows posterior displacement of the anterior mitral-valve leaflet (arrow) and failure of coaptation of the anterior and posterior leaflets (arrowhead). The papillary muscle is echodense and elongated. The view in Panel B (which is rotated 90 degrees from the view in Panel A) shows that the posteromedial papillary muscle (arrowhead) is elongated and that there is prolapse of the anterior mitral-valve leaflet (arrow). LA denotes left atrium, LAA left atrial appendage, and LV left ventricle.

A coronary angiographic study revealed classic aneurysms in left coronary arteries. An abdominal ultrasonographic study showed mild splenomegaly (length of the spleen, 15 cm).

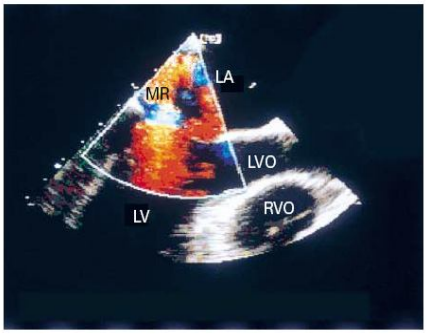

Figure 2. Doppler Color-Flow Transesophageal Echocardiogram.
Severe, posteriorly directed mitral regurgitation (MR) is visible as a band of orange flecked with yellow. RVO denotes right ventricular outflow, LVO left ventricular outflow, LA left atrium, and LV left ventricle.

In the cardiac catheterization laboratory, the patient was in a state of profound cardiogenic shock. Despite the use of an intraaortic balloon pump and a large amount of pharmacologic support, she remained in cardiopulmonary collapse. She was immediately taken to the operating room. A sternotomy was performed to allow exploration. The pericardium and pericardial space were normal. The heart was normal in size but was very hyperdynamic. The great vessels and coronary arteries were of normal distribution and appearance. After cardiopulmonary bypass had been

instituted, the heart was arrested with cold oxygenated blood. The left atrium was normal in size, suggesting that this process was acute. The endocardium of the left atrium looked normal. On first inspection of the mitral valve, the leaflets looked normal, and there were no vegetations. Passive testing with saline showed that the medial aspects of the anterior and posterior mitral valves were regurgitant. On inspection inside the left ventricle, the posteromedial papillary muscle, including its base, was necrotic. The endocardium of the left ventricle, with the exception of the base of the papillary muscle, looked normal. The posteromedial papillary muscle, which had ruptured at the base but was tethered by part of the subvalvular apparatus, was excised. The mitral valve was replaced with a St. Jude Medical prosthesis; the anterolateral papillary muscle and its attachment to the remaining leaflets were preserved. The patient was weaned from cardiopulmonary bypass and taken to the intensive care unit.

On pathological examination the mitral valve was normal, both grossly and microscopically. On gross examination, the papillary muscle was ruptured at its base, and nearly the entire muscle had irregular, sharply demarcated, pale yellow areas with focal hemorrhage-findings consistent with infarction. Histologic evaluation of the papillary muscle showed coagulative necrosis with loss of myocyte nuclei, contraction-band necrosis, and infiltrating neutrophils at the periphery. Within the myocardium, virtually every arteriole was occluded by thrombi composed of fibrin and platelets, a striking and unusual finding.

A Kidney biopsy was performed because of permanent microhematuria and showed neutrophilic infiltration in the wall of kidney medium-sized vessel. A test for antinuclear antibodies was positive (titer, 1:320). Antibodies to double-stranded DNA were absent, but elevated titers of perinuclear antineutrophil cytoplasmic antibodies (P-ANCA) was detected. The C3 and C4 levels were normal. The test for rheumatoid factor was negative and lupus anticoagulants were absent.

DISSCUSION

The most likely diagnosis in this case was systemic lupus erythematosus, a systemic vasculitis such as polyarteritis nodosa or infective endocarditis.[1] Although several features of systemic lupus erythematosus are present, the diagnostic criteria are not met. Also infective endocarditis was probable but the mitral valve was not fully flail. However, the valve leaflets themselves were intact, with no vegetation or other abnormality and the presence of an infarction in the papillary muscle indicate that this patient had a myocardial infarction in the context of an autoimmune disease.[1,2] The infarction could be due either to coronary arteritis or to a coagulopathy; that neutrophilic infiltration in the wall of kidney medium-sized vessels, positive P-ANCA and normal C3 and C4 levels warranted a presumptive diagnosis of microscopic polyarteritis nodosa, even in the absence of other characteristic features.[3]

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